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Identical twins with differing forms of ventricular pre-excitation

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SUMMARY Identical 10-year-old twins, both with electrocardiograms showing a short PR interval and a normal QRS complex but with dramatically different electrophysiological characteristics, are described.

One twin experienced episodes of rapid palpitation and on one occasion was resuscitated from ventricular fibrillation. An intracardiac electrophysiological study confirmed the presence of an atrioventricular nodal bypass tract and in addition revealed the presence of an accessory atrioventricular pathway, thus demonstrating that the patient had both the Lown-Ganong-Levine and Wolff-Parkinson-White syndromes. Re-entry tachycardia and atrial fibrillation, with a very rapid ventricular rate, were precipitated. After treatment with amiodarone, the patient became asymptomatic and a repeat study showed that the features of the atrioventricular nodal bypass tract were no longer present and though re-entry tachycardias using the accessory atrioventricular pathway could still be induced, their rates were slower than before treatment.

The other twin, in spite of an identical surface electrocardiogram, was asymptomatic. An electrophysiological study showed the features of an atrioventricular nodal bypass tract but there was no evidence of additional atrioventricular accessory connections and a tachycardia could not be induced.

The Lown-Ganong-Levine syndrome (1952) and the Wolff-Parkinson-White syndrome (1930) are both types of ventricular pre-excitation. The former results from an atrioventricular nodal bypass tract and the latter from an accessory atrioventricular pathway (Wellens, 1975).

The purpose of this paper is to describe identical twins with differing forms of pre-excitation. Both have the surface and intracardiac electrophysiological features of an atrioventricular nodal bypass tract (Lown-Ganong-Levine syndrome). One twin has in addition, the features of a concealed accessory atrioventricular pathway (Wolff-Parkinson-White syndrome). In cases of ventricular pre-excitation, accessory atrioventricular connections of more than one type are sometimes present but this is only the second report of a patient with the surface electrocardiogram of the Lown-Ganong-Levine syndrome in whom an accessory atrioventricular pathway was disclosed by an intracardiac electrophysiological study (Massumi and Vera, 1971; Spurrell *et al.*, 1973; Gallagher *et al.*, 1976), and it is the first report of identical twins with proved atrioventricular nodal bypass tracts.

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Patients and methods

The patients are identical, male, 10-year-old twins. They were born after an uncomplicated pregnancy and delivery but twin A was noticed to have an apical systolic murmur in infancy and this has persisted. Twin A had experienced frequent, brief episodes of rapid palpitation for some months before the incident which brought about his admission to hospital. At that time his father, a general practitioner, found that he had a heart rate of over 300 per minute; carotid sinus massage was ineffective and 2 hours later, after admission, an electrocardiogram showed atrial fibrillation with an extremely rapid ventricular rate (Fig. 1A). Shortly afterwards he developed ventricular fibrillation and was rapidly defibrillated. Twin B was asymptomatic. Both boys had identical surface electrocardiograms showing a short PR interval, and a normal QRS complex and QT interval (Fig. 1B). Chest radiographs and echocardiograms of both twins were normal as were the electrocardiograms of first degree relatives.

Because of the heart murmur, cardiac catheterisation was performed on twin A and both twins had intracardiac electrophysiological studies under local anaesthesia after premedication with diazepam and

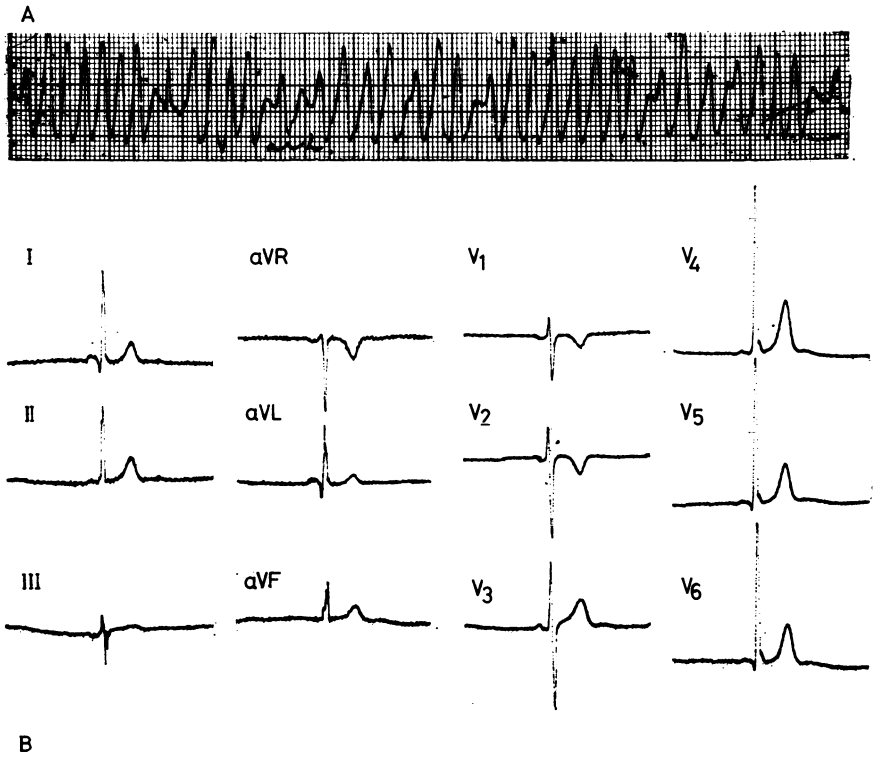

Fig. 1 *(A) Twin A: electrocardiogram on admission showing atrial fibrillation, minimum RR interval 130 ms. (B) The twins had identical electrocardiograms, PR, 100 ms.*

droperidol. At the time of the initial study no other drugs were being taken. Twin A was restudied after 5 weeks' treatment with amiodarone (600 mg daily for 3 weeks and then 400 mg daily). In each study a quadripolar and a tripolar electrode were introduced percutaneously from the right femoral vein. The quadripolar electrode was advanced to the right atrium, the distal poles were used for recording the high right atrial electrogram and the proximal poles for atrial stimulation. The tripolar electrode was positioned across the tricuspid valve to record the low right atrial and His bundle electrograms, and was also advanced into the right ventricle for ventricular stimulation. Two limb leads and lead V1 were also recorded. Recordings were made simultaneously on an ink jet recorder at a paper speed of 100 mm/s. The initial electrophysiological study on twin A was performed after the haemodynamic study and time permitted only a brief examination; for this reason, only atrial and ventricular pacing at varying rates was done, whereas in the study on twin B and in the second study on twin A both rapid pacing and programmed premature stimulation were carried out.

Results

TWIN A

Right and left heart pressures and oxygen saturations, and a left ventricular angiocardiogram were normal; no cause for the systolic murmur was found.

The results of the first electrophysiological study are summarised in Table 1. Atrioventricular nodal

Table 1 *Twin A: conduction times during sinus rhythm and atrial pacing*

	Rate (beats/min)	*AH (ms)*	*HV (ms)*	*QRS complex*
Sinus rhythm	125	50	35	Normal
Atrial pacing	150	50	35	,,
,, ,,	170	50	35	,,
,, ,,	188	60	35	,,
,, ,,	200	60	35	,,
,, ,,	222	60	35	,,
,, ,,	240	70	20	Delta wave
,, ,,	260	70	0	,, ,,
,, ,,	285	His deflection not identified		,, ,,

conduction time (AH interval) was short and increased only minimally with rapid atrial pacing (Fig. 2A) indicating an atrioventricular nodal bypass tract (Castellanos *et al.*, 1971; Caracta *et al.*, 1973; Gallagher *et al.*, 1976). His-Purkinje conduction time (HV interval) was normal. At atrial pacing rates of 240 per minute and greater, the QRS complexes became deformed and the HV interval shortened by the appearance of a delta wave, positive in lead V1 (Fig. 2B) thus showing the presence of either a concealed accessory atrioventricular pathway, or possibly a nodoventricular or fasciculoventricular accessory connection (Mahaim fibres). 1:1 atrioventricular conduction was maintained at the highest rate, 285 per minute, at which the atria were paced. Right ventricular pacing, performed at rates up to 230 per minute, resulted in a constant ventriculoatrial conduction time (VA interval) of 110 ms. Re-entry tachycardia (250 per minute) and atrial fibrillation with virtually all complexes showing type A pre-excitation (minimum interval between pre-excited ventricular complexes 170 ms) each occurred several times during the investigation (Fig. 2C, D).

The results of the second study are summarised in Table 2. Amiodarone prolonged the PR (130 ms),

Table 2 *Twin A after amiodarone: conduction times during sinus rhythm and atrial pacing*

	Rate (beats/min)	*AH (ms)*	*HV (ms)*	*QRS complex*
Sinus rhythm	83	70	35	Normal
Atrial pacing	100	80	35	„
„ „	120	100	35	„
„ „	140	135	10	Delta wave
„ „	150	140	0	„ „
„ „	170	200	−50	„ „
„ „	188	205	−55	„ „
„ „	200	210	−60	„ „
„ „	210	230	−70	„ „
„ „	220	2:1 atrial block		

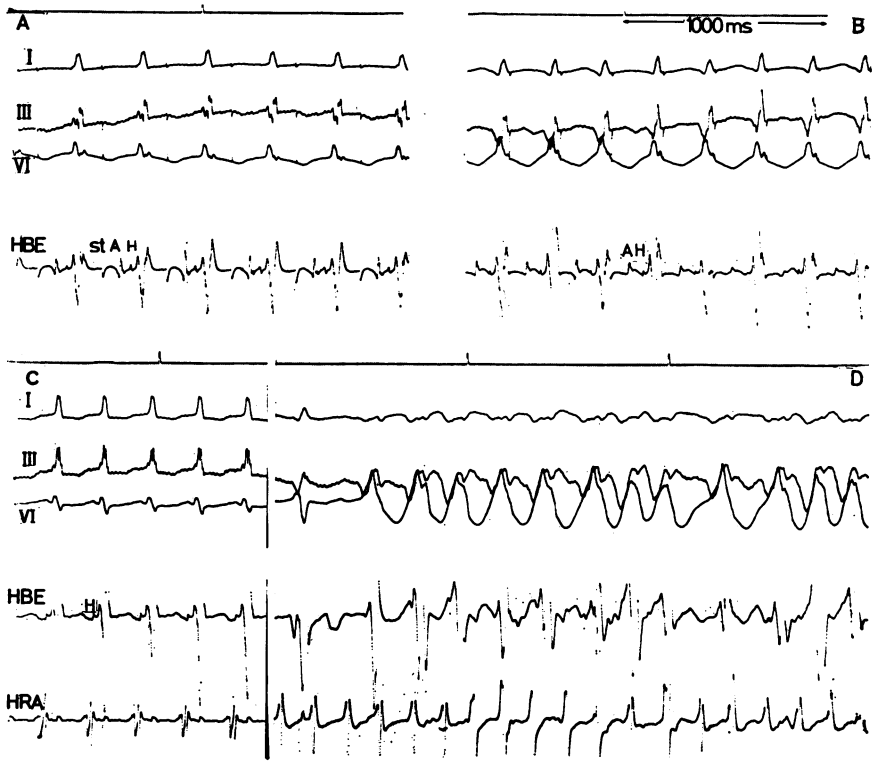

Fig. 2 *Twin A: (A) atrial pacing at 188 per minute, AH, 60 ms, note the distinct atrial latency period; (B) atrial pacing at 240 per minute led to the appearance of delta waves, AH, 70 ms; (C) re-entry tachycardia, rate 250 per minute; (D) atrial fibrillation, minimum RR interval 170 ms, conduction mainly via accessory atrioventricular pathway. HBE and HRA, His bundle and high right atrial electrograms, respectively; st, pacing stimulus; A and H, atrial and His bundle depolarisation respectively.*

AH (70 ms) and QT (QT_c 420 ms) intervals. In contrast to the first study, distinct increases in the AH interval occurred with rapid atrial pacing and with single premature atrial stimuli. Thus, the features of an atrioventricular nodal bypass tract were no longer present. However, atrial pacing at rates of 140 per minute and greater, and single atrial stimuli within 440 ms of the preceding atrial beat led to the appearance of a delta wave (Fig. 3A). As the cycle length was shortened, the delta waves, which had similar vectors to those seen in the first study, became progressively larger and the HV interval became progressively shorter. These are the features of an accessory atrioventricular pathway rather than Mahaim fibres (Wellens, 1975; Gallagher *et al.*, 1976). Re-entry tachycardia using the accessory atrioventricular pathway in both anterograde (225 per minute) and retrograde (167 per minute) directions occurred but their rates were considerably less than found in the first study and atrial fibrillation did not occur (Fig. 3B, C). Twin A has been asymptomatic in the 8 months since amiodarone was started.

TWIN B

The results are summarised in Table 3. During sinus rhythm values for the AH and HV intervals were the same as in Twin A. On rapid atrial pacing and with single premature atrial stimuli small increases in AH interval occurred, but even at 230 per minute, the maximum rate at which 1:1 AV conduction was maintained, the AH interval had increased by only 65 ms. Even at the shortest cycle length delta waves did not appear. Right ventricular pacing up to 200 per minute resulted in a constant VA interval of 110 ms; above this rate 2:1 ventriculo-atrial block occurred. These findings indicate an atrioventricular nodal bypass tract but there was no

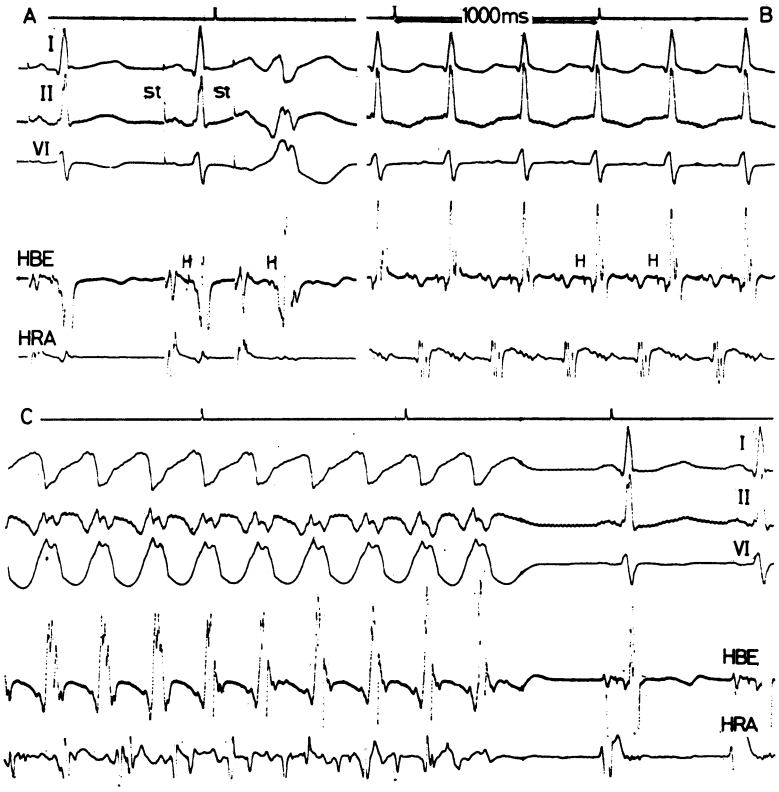

Fig. 3 *Twin A after amiodarone: (A) premature atrial stimulus at 340 ms prolonged AH to 150 ms and produced delta wave; (B) re-entry tachycardia, rate 167 per minute, using accessory atrioventricular pathway in retrograde direction; (C) re-entry tachycardia, rate 225 per minute, using accessory atrioventricular pathway in anterograde direction, spontaneously reverting to sinus rhythm.*

Table 3 *Twin B: conduction times during sinus rhythm and atrial pacing*

	Rate (beats/min)	*AH (ms)*	*HV (ms)*	*QRS complex*
Sinus rhythm	103	50	35	Normal
Atrial pacing	110	50	35	,,
,, ,,	130	60	35	,,
,, ,,	150	80	35	,,
,, ,,	170	90	35	,,
,, ,,	190	95	35	,,
,, ,,	200	100	35	,,
,, ,,	220	110	35	,,
,, ,,	230	115	35	,,
,, ,,	240	115	2:1 infra Hisian block	
,, ,,	250	2:1 supra Hisian block		

evidence of an accessory atrioventricular pathway. A tachycardia could not be precipitated by either premature atrial or ventricular stimulation.

Discussion

It has been known for many years that the Wolff-Parkinson-White syndrome can occur in several members of a family, and it has been reported in a set of identical twins (Wolff *et al.*, 1930; Oehnell, 1944; McIntire and Freed, 1955; Harnischfeger, 1959; Massumi, 1967; Schneider, 1969). There are, however, only 2 reports of a familial incidence of the Lown-Ganong-Levine syndrome. Lown *et al.* (1952) mentioned a mother and son with the disorder and recently Brodsky *et al.* (1977) described 6 members of a family with the surface electrocardiographic features of the Lown-Ganong-Levine syndrome though the proband was studied by intracardiac electrical stimulation and found not to have the features of an atrioventricular nodal bypass tract. The present report is the first of identical twins each with electrophysiological evidence of an atrioventricular nodal bypass tract. In theory, patients with an atrioventricular nodal bypass tract should show no increase in AH interval with premature atrial stimulation. In practice, however, this is a rare finding and usually, as in our patients, small increases in AH interval do occur (Castellanos *et al.*, 1971; Caracta *et al.*, 1973; Wellens, 1975). The increase in AH interval in Twin B was greater than in Twin A and the former might be considered to have a 'partial' rather than 'complete' bypass tract. It should be pointed out that though the electrophysiological characteristics of an atrioventricular nodal bypass tract are well recognised there is doubt as to whether the tract is extranodal or intranodal and whether it is a functional rather than an anatomical entity (Wellens, 1975). Because of these doubts Gallagher *et al.* (1976) prefer to use the term 'enhanced AV node conduction'.

A further point of interest was the differing forms of pre-excitation in the twins; Twin A having an accessory atrioventricular pathway in addition to an atrioventricular nodal bypass tract. Presence of the accessory atrioventricular pathway was concealed during sinus rhythm and it required very short cycle lengths to cause slight but sufficient prolongation of atrioventricular nodal bypass tract conduction so that it was slower than conduction along the accessory atrioventricular pathway and hence led to the appearance of delta waves. A similar case has been reported by Massumi and Vera (1971). One possibility to be considered is that the delta waves were the result of Mahaim fibres since an accessory atrioventricular pathway can be mimicked by a combination of an atrioventricular nodal bypass tract and Mahaim fibres (Brechenmacher *et al.*, 1976). However, the normal HV interval during sinus rhythm, and the progressive increase in degree of pre-excitation and progressive shortening of the HV interval with decreasing cycle lengths (seen particularly in the second study) are against this possibility (Wellens, 1975; Gallagher *et al.*, 1976). Nevertheless, as pointed out by Wellens (1975), Mahaim fibres inserted into the atrioventricular node in such a location as to bypass much of the part of the node responsible for causing delay in atrioventricular conduction might be indistinguishable from an accessory atrioventricular pathway. The demonstration of left atrial pre-excitation during ventriculoatrial conduction by recording a left atrial electrogram during ventricular pacing would have been helpful in supporting the diagnosis of an accessory atrioventricular pathway, and also in confirming that the pathway was left sided, as suggested by the type A delta waves (Wellens, 1975).

In Twin B no evidence of an accessory atrioventricular pathway was found. It is possible that this pathway was present but was only able to conduct retrogradely. Here again confirmation of this possibility would have required a demonstration of left atrial pre-excitation during ventriculoatrial conduction but even if an accessory atrioventricular pathway had been present ventriculoatrial conduction via the atrioventricular nodal bypass tract might have prevented left atrial pre-excitation.

In summary, identical twins with the same surface electrocardiographic signs of pre-excitation were found to have very different electrophysiological characteristics.

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